230* Intravenous methylprednisolone pulse therapy vs. oral prednisone for allergic bronchopulmonary aspergillosis (ABPA) in CF

M. Cohen-Cymberknoh[1], H. Blau[2], D. Shoseyov[1], O. Efrati[3], S. Armoni[1], E. Kerem[1]. [1]*CF Center, Hadassah University Hospital, Jerusalem, Israel;* [2]*Schneider Children's Medical Center, Petah Tikva, Israel;* [3]*Sheba Medical Center, Tel Hashomer, Israel*

The treatment of ABPA consists of prolonged oral corticosteroids and antifungal therapy which may have severe toxicity. High-dose IV pulse methylprednisolone therapy (HDIVPMT) is used for autoimmune diseases and was shown to be as effective as oral prednisone and associated with less side effects.
The purpose of this study was to compare treatment with HDIVPMT to conventional therapy with oral prednisone in patients with CF suffering from ABPA. Nine patients (5 females) ages 7–36 years old were treated with HDIVPMT (10 mg/kg/d) for 3 days once a month, and 5 patients received oral prednisone (1–2 mg/kg/d), until clinical and laboratory resolution of ABPA signs was noted. All the 14 patients were also treated with itraconazole.
Results: Both groups showed similar clinical and laboratory improvement (increase in FEV_1, decline in serum IgE levels (total and specific anti AF), and total eosinophil counts. The adverse effects in the group that received HDIVPMT were minor and included flushing, myalgia and restlessness, all of which disappeared within 3–5 days after the pulse. The duration of therapy was 4–12 pulses. Patients that received oral prednisone suffered from more serious side effects including cushingoid facies, hirsutism, severe weight gain, depression, emotional instability and hypertension, all were reversible only after steroid dose was reduced or stopped. The duration of oral therapy was much longer and more patients needed repeated therapies.
In conclusion, pulse methylprednisolone is as effective as oral prednisone with less serious side effects and therefore is advantageous in the treatment of ABPA in CF.

231 21% prevalence of ABPA in a regional paediatric Cystic Fibrosis (CF) clinic – how high is too high?

C.A. Byrnes[1,2], M. Jaksic[1], J.C. Tate[1]. [1]*Starship Children's Hospital, Auckland, New Zealand;* [2]*University of Auckland, Auckland, New Zealand*

Aim: To document the prevalence, severity and response to treatment of ABPA in children with CF followed up in a single, regional clinic.
Method: Retrospective review of all APBA diagnoses from 2000 to 2007 inclusive in children 0–15 years of age at the CF Clinic, Starship Children's Hospital, New Zealand.
Results: There were 32 episodes of ABPA seen in 16 children (mean age 10 years, range 4–15 years, 12 males) in the clinic population of 75 giving a prevalence of 21%. The mean total IgE was 2248 kIU/L (range 174–10,300) and >1000 kIU/L in 23 episodes at time of diagnosis. The eosinophil counts were raised in 9 episodes, all had positive RAST testing and aspergillus precipitans with 15/16 growing aspergillus in sputum. FVC decreased by 22.1% predicted (range 0–65%) and FEV1 by 23.9% predicted (range 0–70%) during the acute event. There were no significant differences between FVC and FEV1 from last best prior to ABPA episode and post ABPA treatment. Nine were hospitalised, 2 required intensive care and 3 needed oxygen. All were treated with oral steroid therapy with mean duration 8.9 months (range 0.5–31 months) and all had oral intraconazole. Novel treatments were tried because of poor response; 3 children had 4 courses of methylprednisolone and 3 had nebulised amphotericin. The incidence increased from 3–4 episodes/year to 8 in 1 year on completion of building works within the hospital.
Discussion: A prevalence of 21% ABPA is higher than previous reports. The disease described is severe and refractory to treatment. Surprisingly, there was no significant difference in lung function testing before and after the ABPA episodes. Recent hospital construction and the high background rate of atopy in NZ children may be factors.

232 Infection with *Mycobacterium abscessus* deteriorates lung function dramatically in CF patients

K. Kötz[1], M. Gilljam[1], A. Lindblad[1]. [1]*The West Swedish CF Centre, Queen Silvia Children's hospital, Gothenburg, Sweden*

Objective: To assess the clinical course in patients with chronic *M. abscessus* infection.
Patients: At our centre 5/140 CF patients are chronically infected with *M. abscessus*. All are PI and were between 11 and 25 years old at the time of infection. Two patients were chronically infected with *P. aeruginosa*, one with *B. cepacia*, one with *S. maltophilia* and one patient had no other chronic infection. All were treated with at least three antibiotics and/or IFN-γ for 10 till 24 months. Only patient 4 was BCG-vaccinated as a child.
Results: FEV_1 decreased dramatically in 3 patients: 32 and 43% within 7 resp. 41% within 10 years. Two patients developed hearing impairment due to long-term treatment with amikacin. The fourth patient had an average annual loss of 0.5% in FEV_1%. The fifth patient is infected with *M. abscessus* since $2\frac{1}{2}$ years. His FEV_1 deteriorated severely before detection of *M. abscessus* but improved after the start of antibiotic/immunologic treatment.
Conclusions: Three of 5 patients had a long-term decrease of FEV_1% despite aggressive therapy causing severe hearing impairment in two patients. A multicenter-study is warranted to evaluate the most effective treatment.

Patient	FEV_1%					
	1 yr before infection	When infected	2 yrs after infection	7 yrs after infection	10 yrs after infection	Annual ΔFEV_1%
♀ 17 yrs	122	114	113	83	73	–4.1
♀ $12\frac{1}{2}$ yrs	114	115	100	83		–4.6
♂ $11\frac{1}{2}$ yrs	66	62	51	19		–6.1
♂ $25\frac{1}{2}$ yrs	86	81	81	81	76	–0.5
♂ $11\frac{1}{2}$ yrs	104	88	97			+4.5

233 Clinical difference between cystic fibrosis patients colonized with different bacteria

P. Reis[1], J. Fermeiro[1], S. Castanhinha[1], T. Rodrigues[2], L. Pereira[1], C. Barreto[1]. [1]*Cystic Fibrosis Paediatric Centre, Santa Maria Hospital, Lisbon, Portugal;* [2]*Biomathematics Laboratory, Lisbon, Portugal*

Introduction: The aim of our study is to analyse the clinical difference between CF patients (pts) with *Staphylococcus aureus* (SA), *Pseudomonas aeruginosa* (PA) and *Burkholderia cepacia* (BC) chronic colonization.
Methods: Retrospective review of clinical charts and bacterial isolation of all the pts followed in 2007. Number of hospitalizations, number of days in hospital and respiratory function tests (RFT) – FEV1; FEF 25–75 and FVC, were evaluated. Data were analysed with SPSS 14.0 version, using T-test, two sample, and Man-Whitney test.
Results: 56 pts were included (55% female; median: 9 years). SA, PA and BC chronic colonization were found respectively in 41.1% (23/56), 23.2% (13/56) and 4.1% (4/56) of the pts. Pts with no chronic colonization had better results on RFT, but the difference was not significant (p value >5%). Comparing pts with different bacteria chronic colonization with those who did not have that specific colonization, we found that PA and BC colonised pts had worse RFT results. Pts who did not have PA chronic colonization had better FEV1 (95% CI 3.6–40.1), FEF 25–75 (95% CI 0.5–51.8) and FVC (95% CI 0.3–32.2). Pts with no BC chronic colonization had higher FEV1 (95% CI 16.5–74.6) and FVC (95% CI (25.9–71.5). There was no statistical significant difference regarding RFT results and SA chronic colonization. BC colonized pts had higher number of hospitalizations and number of days in hospital (p: 0.014; p: 0.012). That was not found in the other colonized groups.
Discussion: PTS with PA and BC chronic colonization had worse RFT. This was not found in SA cases. BC chronic colonization is associated with a higher number of hospitalizations that last longer.
Special Thanks: SMH Microbiology Department.